

## What is a vascular malformation?

A vascular malformation is an abnormal growth of blood vessels or lymph vessels that occurs before birth. Unlike hemangiomas, they grow at a steady rate and do not rapidly increase in size. Also unlike hemangiomas, these do not go away on their own or go away without treatment.

Vascular malformations affect boys and girls at the same rate and although present at birth, may not be detected.

A second and completely distinct type of vascular lesion present is a vascular malformation. A vascular malformation is any sort of abnormal blood vessel or lymph vessel growth that occurs in a patient that is not related to hemangioma. These lesions result from an abnormal development of vessels before birth. They usually grow at a normal rate throughout life and they do not often rapidly increase in size. However, they also do not go away without treatment. Vascular malformations can affect boys and girls at same frequency; usually they are present at birth, although some can go undetected. It is essential to separate a vascular anomaly from a hemangioma, as their treatments are markedly different. Vascular anomalies do not respond to steroids, interferon, or anticancer drugs such as vincristine. They can be treated immediately at birth if they will not go away and do require multiple treatments over the lifetime.

The most common vascular anomaly seen is a capillary malformation called a port-wine stain. This is sometimes called an angel kiss or stork bite. It can be pink, red, purple, or a variety of discolorations that are usually on the surface of the skin and are usually not raised. They are made up of small, teeny, tiny blood vessels called capillaries, which are just behind the skin. They frequently are on the face but can be seen in other parts of the body. One kind of port-wine stain, which is important to be aware of is Sturge-Weber syndrome. This facial port-wine stain can involve the brain and can, over the long term, produce glaucoma or seizures. It needs to be evaluated very closely by a pediatric plastic surgeon as well as a neurologist.

Laser treatments are excellent for vascular anomalies such as port-wine stains and other capillary malformations that are less effective in treating

hemangiomas. Lasers are typically reserved in hemangiomas for those that are bleeding or ulcerating early during the proliferative phase or following the involution phase for treating small telangiectasias or abnormal blood vessels. It is not realistic to think that a laser will make a proliferating hemangioma go away. In terms of vascular birthmarks, treatment needs to be repeated multiple times in order to maximize the removal of the lesion and prevent any burning or scarring on the skin. Many of these lesions can also be covered with topical cosmetics and working with a good cosmetologist can significantly improve the appearance of many of the lesions, especially the flatter ones on the face.

Another common type of vascular malformation is the lymphatic malformation. These have also been called cystic hygromas. These anomalies occur following abnormal growth of lymph vessels, which carry fluid from our lymph glands through out blood and help remove bacteria from our body tissues. Lymph vessels drain fluid from areas of the body that become swollen after infection. If they are malformed at birth, they often look like large, soft masses and are usually in the head and neck region. They are often tangled all around muscles, skin, and bones.

Infection is the most common problem associated with lymphatic malformations or lymphangioma. While most infections are minor and disappear with antibiotics, some are very severe, causing rapid growth and must be treated surgically. Because repeat infections are common, your doctor may recommend prophylactic treatment not only with antibiotics, but also by surgical and interventional radiology methods.

Dr. Stelnicki frequently works with a pediatric radiologist to optimize the treatment of these lesions. He may order an MRI (magnetic resonance imaging) study and a special blood vessel study to evaluate the lymphangioma or vascular malformation prior to any therapy. Prior to surgery he will consult the interventional radiology unit at for either percutaneous sclerotherapy and intravascular embolization. This decreases the size of the lesion and the amount of bleeding. It is our practice to involve a multidisciplinary approach to decreasing these lesions so as to decrease the amount of surgeries needed and the amount of overall scarring

Heart problems can also occur with vascular birthmarks called arterial venous malformations. These malformations can steal blood flow away from important systems in the body and cause the heart to work harder than it should and cause heart failure. If this occurs, then early intervention is required

Vascular malformation, surgery is reserved for those large lesions that are causing overgrowth of an organ system in an area significant distorting tissue and all surgery needs to be performed by a combination of both interventional radiology techniques and direct surgical excision. Dr's Stelnicki works hard to decrease the amount the of scarring that will be present following any surgical interaction.

It is important to be involved not only in the vascular anomalies team but also develop a support network. Dr. Stelnicki, through the Facing it Together Foundation has linked families of children with vascular anomalies. This allows families to realize they are not the only ones dealing with this problem. Our office will help you contact other families who are going through the same problem and have also been through it, so that you can have some point of reference as to what to expect in the upcoming years. Dr. Stelnicki and the vascular anomalies team at Joe DiMaggio Children's Hospital will follow you closely throughout your child's life until the treatment for your hemangioma or vascular malformation has been optimized. Additional information is also available through the Cleft Palate Foundation that has a link on this web site.

### What is a port wine stain?

The most common vascular anomaly in this category is a capillary malformation called a port wine stain. This is sometimes called an angel kiss or stork bite. It can be pink, red, purple, or a variety of discolorations that are usually on the surface of the skin and not raised. They are comprised of tiny blood vessels called capillaries.

Frequently they are on the face, but may be seen on other parts of the body.

### What is Sturge-Weber syndrome?

Sturge-Weber syndrome is a type of port wine stain that may involve the brain and in the long-term, cause glaucoma, seizures, intracranial vascular lesions, and leptomeningeal calcifications. It involves the capillaries along the maxillary and mandibular branches of the trigeminal nerve. This type is important to be evaluated by a pediatric plastic surgeon, neurologist, and ophthalmologist.

### What is a lymphatic malformation?

A lymphatic malformation (lymphangioma) is also commonly called a cystic hygroma. These occur with abnormal growth of lymph vessels. The lymph system carries fluid from lymph glands and help remove bacteria. When they are malformed at birth, they look appear as large, soft masses usually located in the head and neck region. They are often intertwined around muscles, skin and bone.

Infection is commonly seen with lymphatic malformations and is usually addressed with antibiotics. In the case of a severe infection and large, rapid growth, these must be treated surgically. The doctor may recommend prophylactic antibiotics and surgical and interventional radiologic treatment methods.

### What is an arteriovenous malformation?

An arteriovenous malformation is composed of abnormal connections between the arteries and veins. These malformations can steal blood flow away from important systems in the body, causing the heart to work harder than it should and thus putting the patient at the risk of heart failure.

### What is a venous malformation?

A venous malformation is composed of enlarged sponge-like veins. Treatment is indicated if they cause pain, distortion, obstruction, or bleeding.

### What are the treatments for a vascular malformation?

Dr. Stelnicki may order an MRI scan or special blood vessel study to evaluate the AVM or lymphangioma.

Vascular laser treatments are excellent for treating vascular malformations.

Because these grow throughout life, they do require multiple treatments with the laser in a lifetime.

If surgery is warranted, Dr. Stelnicki may consult interventional radiology for either percutaneous sclerotherapy or intravascular embolization. These methods help reduce the amount of bleeding and scarring. Surgery is reserved for large lesions that are causing overgrowth of an organ system and distorting tissue.